CASE REPORTS

Ischemic intestinal involvement in a patient with Buerger disease: Case report and literature review

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A 42-year-old Japanese man who had undergone amputation of the left leg below the knee because of Buerger disease required emergency thrombectomy 7 months later. He complained of acute abdominal pain after thrombectomy. At aortography the distal superior mesenteric artery and its branches were not well visualized. Emergency laparotomy was performed because of suspected intestinal ischemia, and the terminal ileum and cecum and part of the ascending colon were resected. In total, the patient underwent laparotomy four times. Histopathologic findings revealed that the arteries and veins of the resected small intestine were occluded with organized thrombi. Inflammatory cell infiltration was recognized mainly in the intima. These findings are compatible with Buerger disease. (J Vasc Surg 2003;38:170-4.)

Buerger disease was described and established in the English literature in 1908[1] as a clinicopathologic entity distinct from atherosclerosis. Today Buerger disease is accepted as a definite vascular disease with typical clinical presentation, natural history, and histopathologic features.[2-6] The disease usually affects medium and small arteries and veins of the upper and lower extremities. Visceral artery involvement is rarely reported. We report a case of Buerger disease with intestinal ischemia caused by superior mesenteric artery (SMA) occlusion. In addition, review of the literature to date is presented.

CASE REPORT

A 42-year-old Japanese man was admitted to our department with numbness and pain in the left calf in March 2000. Onset of symptoms had begun 7 days earlier, and he treated left calf pain himself with a commercial painkiller. When seen by his local physician, magnetic resonance angiography was performed, which revealed occlusion of the left popliteal artery. The patient was brought to our university hospital by ambulance.

At admission to the emergency room, the left lower limb pulses were not palpable except for the femoral artery; however, pulses in the upper extremities all were palpable. Soon after admission the patient underwent emergent thrombectomy of the left leg, and continuous intra-arterial infusion of prostaglandin E was administered through the left common femoral artery. However, it became necessary to amputate the left leg below the knee 1 month

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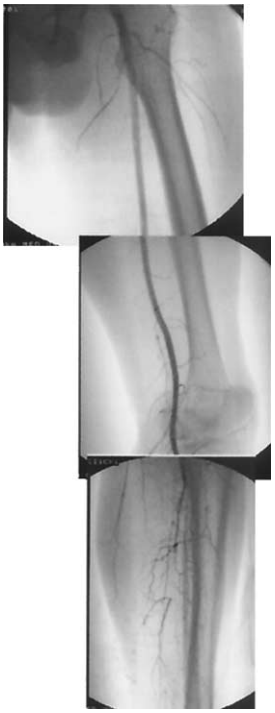

Fig 1. Postoperatvive angiogram obtained just after removal of continuous intra-arterial infusion catheter. Right superficial femoral artery and popliteal artery are both patent, and distal site of occluded popliteal artery has well-developed collateral vessels.

Table I. Clinical criteria[4] for diagnosis of Buerger disease*

Smoking history
Onset of symptoms before age 50 years
Infrapopliteal arterial occlusive lesions
Either upper limb involvement of phlebitis migrans
Absence of atherosclerotic risk factors other than smoking

*Clinical diagnosis of Buerger disease is made only when all five requirements are met.

Fig 2. Aortogram obtained 14 hours after thrombectomy to treat a second event shows left iliac artery is occluded again. Superior mesenteric artery has a tapering occlusion, and distal arteries are not visualized.

after the first revascularization (Fig 1). The patient had smoked about 40 cigarettes per day for 20 years, but had no other risk factors for atherosclerotic disease, eg, hypertension, hyperlipidemia, diabetes mellitus, although lower limb phlebitis migrans was present.

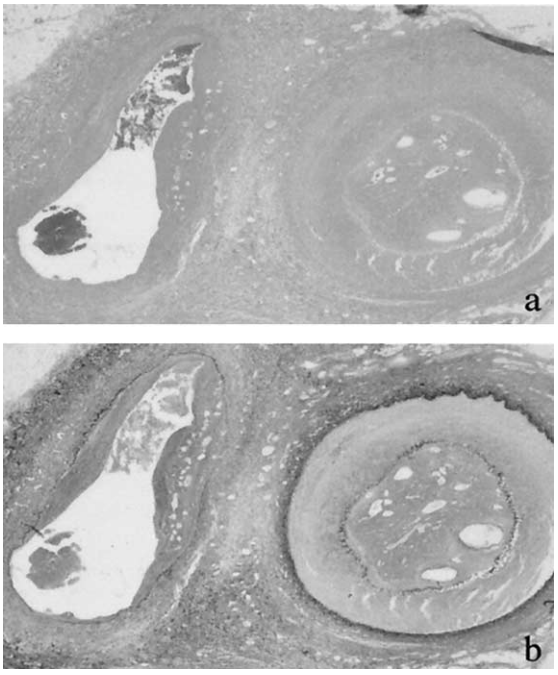

Fig 3. Posterior tibial artery contains organized thrombus, and recanalization is noted. All three layers are almost intact, and inflammatory cell infiltration is detected, mainly in intima and media. Adjacent posterior tibial vein has fresh thrombus and thickening intima. Inflammation is recognized in the intima. **a,** Hematoxylin-eosin, original magnification × 200. **b,** EVG, original magnification × 200.

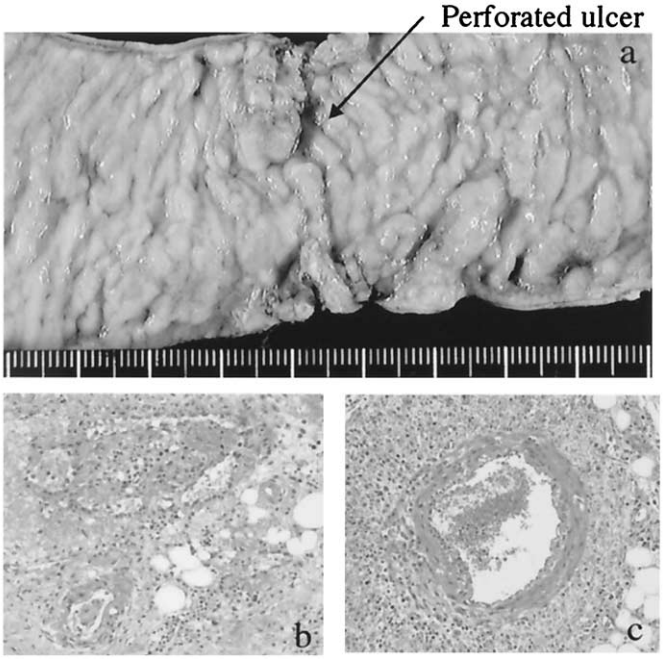

Fig 4. Macroscopic spicemen of resected ileum. a, Perforated ulcer (arrow) and some nonperforated ulcers are recognized. b, c, Marginal artery and vein have fresh thrombi and thickening and inflamed intima.

Table II. Reported cases of visceral intestional Buerger disease

Case	*Author*	*Sex*	*Age (y)*	*Symptom*	*Affected artery*	*Treatment*
1	Wolf and Marshak[9]M	M	40	Bowel obstruction	Jejunoileal	Small bowel resection
2	Rob[10]	M	42	Abdominal pain, weight loss	Small intestinal	Bowel resection
3	Herrington and Grossman[11]	M	33	Acute abdominal pain	Sigmoid	Sigmoidectomy
4	Herrington and Grossman[11]	M	42	Abdominal pain, weight loss	Jejunal	Bowel resection
5	Cbezas-Moya and Dgragstedt[12]	M	33	Bowel obstruction	Jejunal	Bowel resection
6	Wolf et al[13]	M	53	Abdominal pain	Jejunal	Bowel resection
7	Horie et al[14]	M	43	Shock	Jejunal	Conservative
8	Guay et al[15]	M	33	Abdominal pain	Mesocolon	Colectomy
9	Sachs et al[16]	M	45	Abdominal pain	Mesocolon	Bowel resection
10	Borlaza et al[17]	M	36	Abdominal pain	Jejunal	Bowel resection
11	Deitch and Sikkema[18]	M	42	Abdominal pain	Superior mesenteric	Bowel resection
12	Soo et al[19]	M	48	Abdominal pain	Sigmoid	Bowel resection
13	Rosen et al[20]	M	26	Abdominal pain	Colonic	Bowel resection
14	Iyer and Mait[21]	M	39	Abdominal pain	Rectal	Abdominoperineal resection
15	Broid et al[22]	M	20	Abdominal pain	Superior mesenteric	Bowel resection
16	Ito et al[23]	M	42	Abdominal pain	Ileocecal	Bowel resection
17	Schellong et al[24]	M	23	Abdominal angina	Celiac, superior mesenteric	Bypass
18	Sauvaget et al[25]	M	32	Abdominal pain, weight loss	Sigmoid	Sigmodiectomy
19	Lie[26]	M	37	Abdominal pain	Ileocecal	Bowel resection
20	Lie[26]	M	41	Abdominal pain	Colonic	Bowel resection
21	Lie[26]	F	38	Abdominal pain	Jejunal	Conservative
22	Lie[26]	F	35	Abdominal pain	Sigmoid	Conservative
23	Iwai[27]	M	51	Epigastralgia	Right gastric	Gastrectomy
24	Iwai[27]	M	43	Abdominal angina	Celiac, superior mesenteric, inferior mesenteric	Bypass
25	Iwai[27]	M	43	Abdominal angina	Celiac, superior mesenteric, inferior mesenteric	Bypass
26	Our case	M	46	Abdominal pain	Superior mesenteric, inferior mesenteric	Bowel resection

Neither collagen disease nor cardiac disease had ever been diagnosed in this patient. This was the first time a diagnosis of Buerger disease was made, according to our criteria (Table I). The patient stopped smoking only during hospitalization, and he was discharged with daily oral anticoagulant medicine, warfarin sodium and ticlopidine.

Seven months later he returned to our department with acute arterial thrombosis in the bilateral lower extremities. Bilateral femoral artery pulses were hardly palpable, and the lower popliteal artery pulses were not palpable at all. Symptoms of ischemia had persisted for more than 24 hours. Laboratory data included serum creatinine phosphokinase concentration 995 U/mL, and emergent thrombectomy was performed to avert bilateral hip joint amputation. After thrombectomy because of bilateral arterial occlusion, the right popliteal artery and its trifurcation and the left distal popliteal artery were evident on an intraoperative arteriogram. Vascular status in both lower limbs worsened, and 14 hours after thrombectomy, angiography was performed through the right brachial artery with the Seldinger method to confirm whether the lower limb arteries were patent. The distal left iliac arteries and bilateral internal iliac arteries were not visualized, and the peripheral branches of the superior mesenteric artery were not well visualized (Fig 2).

Two days after thrombectomy the patient complained of abdominal pain, with muscle guarding. Emergency laparotomy was judged necessary because of suspicion of ischemic intestinal

Table III. Clinical characteristics of reported cases

Case	*Raynaud phenomenon*	*Phlebitis migrans*	*Tobacco use*
1	No	No	No
2	No	Yes	No
3	Yes	Yes	Yes
4	Yes	Yes	Yes
5	Yes	Yes	Yes
6	Yes	Yes	Yes
7	No	No	Yes
8	No	Yes	Yes
9	No	No	Yes
10	Yes	Yes	Yes
11	No	Yes	Yes
12	No	No	Yes
13	No	Yes	Yes
14	No	No	Yes
15	Yes	No	Yes
16	Unknown	Unknown	Yes
17	No	No	Yes
18	No	No	Yes
19	Unknown	Unknown	Unknown
20	Unknown	Unknown	Unknown
21	Unknown	Unknown	Unknown
22	Unknown	Unknown	Unknown
23	No	No	Yes
24	Yes	No	Unknown
25	No	No	Unknown
26	No	Yes	Yes

involvement. At laparotomy, the ileum end, cecum, and proximal side of the ascending colon and sigmoid colon were necrotic. The necrotic small and large intestines were resected, and a stoma was created. At 10 and 14 days after the first operation, residual intestinal ischemia progressed, and additional intestinal resection and stoma placement were performed. The postoperative course was complicated by recurrent intestinal ischemic perforation, sepsis, and renal and liver dysfunction. After 42 days the patient died of multiple organ failure.

Autopsy findings revealed that the posterior tibial artery was affected, compatible with Buerger disease. Pathologic examination revealed that the posterior tibial artery had intact elastic lamina, with organized thrombi, and three layers preserved almost intact. Inflammatory cell infiltration was detected in the intima and media. The adjacent posterior tibial vein had scarce fresh mural thrombi, with intimal hyperplasia. Adventitial inflammatory infiltration cells were not recognized in either vein in any of the three layers (Fig 3). Resected specimens of small intestine demonstrated ischemic ulcers, and capillaries of the ischemic intestine exhibited thickening of the intima and fresh thrombus formation, with some inflammatory cells. The marginal arteries and veins had well-preserved architecture (Fig 4).

DISCUSSION

In 1908 Buerger described the first case of thromboangiitis obliterans (Buergers disease) and reported that this obliterating vasculopathy could involve the visceral arteries.[1] However, this uncommon vasculopathy mainly affected medium and small arteries in the extremities, and visceral and cerebral arteries were rarely affected.[1-4] Our patient had rare intestinal ischemia caused by Buerger disease. The patient had typical symptoms of Buerger disease at the first visit. He had no atherosclerotic risk factors except for tobacco abuse, and no serologic data suggested any collagen disease or anticoagulation disorder. An electrocardiogram revealed no atrial fibrillation that could cause distal embolization. An angiogram of the lower limb showed findings typical of Buerger disease, eg, abrupt occlusion and tree root pattern. No typical pattern of Buerger disease was detected on an abdominal aortogram, because selective superior mesenteric arteriography could not be performed because of acute renal failure. However, pathologic examination demonstrated that the arteries of the resected small intestine had organized thrombi, with an essentially intact elastic lamina and inflammatory cell infiltration.

These findings are compatible with pathologic findings reported in the literature.[2,4,7] Including our patient, only 26 cases of visceral Buerger disease have been reported to date.[8] We reviewed the clinical characteristics of 26 cases[9-27] (Tables II and III). In these 26 cases, mean (ÅSD) age of patients with visceral Buerger disease was 39.1 Å 8.07 years, and only 2 patients (7.4%) were women.[26] Abdominal symptoms were present in all but 2 patients.[11-14] Symptoms were chronic in 6 patients[17,26,27] and acute in the others. Preoperatively or postoperatively, 6 patients with acute symptoms of Buerger disease had shock, and they died in hospital. Twenty of the 26 patients underwent digestive organ resection; however, in 1 patient the ischemic bowel could not be resected.[14] Small intestinal arteries, ie, superior mesenteric, jejunal, ileocecal, and marginal arteries, were resected in 20 patients, and large intestinal arteries in 10 patients.

Intestinal Buerger disease demonstrates the same histopathologic findings as in arterial and venous lesions. Whereas intestinal Buerger disease is rare, ischemia of the upper or lower extremities is the most common clinical manifestation. In our patient, other vasculopathies could be considered in the differential diagnosis. Some collagen diseases, eg, lupus erythematosus, which induces anticardiolipin antibodies, can cause thromboembolic manifestations.[28] Polyarteritis nodosa affects small and medium muscular arteries in many organs. In these diseases, the gastrointestinal tract is commonly involved. Bleeding and perforation are not rare manifestations. Mortality ranges between 75% and 100%.[29]

In our patient, collagen disease was ruled out because no autoimmune antibodies or anticardiolipin antibodies were evident at serologic testing. Furthermore, histopathologically affected arteries demonstrated an essentially intact internal elastic lamina, and all three layers were well preserved, with intimal cell infiltration. These findings are strongly characteristic of Buerger disease. Furthermore, there was no fibrinoid necrosis, no cell infiltration through all three layers, and no complete destruction or fragmentation of the internal elastic lamina.

Perioperative mortality was, surprisingly, only 30%, despite emergency surgery. Although intestinal Buerger

disease is rare, once the digestive tract is affected it becomes a life- threatening disease. During the course of this disease intestinal involvement is possible, and careful observation for abdominal symptoms is mandatory.

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